Pemphigus is a group of rare but potentially fatal diseases. Pemphigus is an autoimmune disease with clinical manifestations of chronic intraepidermal bullae on the skin and mucosa, and loose walls. Pemphigus can be categorized into four types, namely pemphigus vulgaris (PV), pemphigus vegetans, pemphigus foliaceus, and paraneoplastic pemphigus. Pemphigus vulgaris is the most common form of pemphigus with the highest epidemiology. Pemphigus vulgaris with abdominal tumors can not be associated where abdominal tumors do not trigger the occurrence of PV. However, abdominal tumors can increase morbidity and provide poor prognosis.

**Background:** Pemphigus is an autoimmune disease with clinical manifestations of chronic intraepidermal bullae on the skin, mucosa, and loose walls. Pemphigus can be categorized into four types, namely pemphigus vulgaris (PV), pemphigus vegetans, pemphigus foliaceus, and paraneoplastic pemphigus. Pemphigus vulgaris is the most common form of pemphigus with the highest epidemiology. Pemphigus vulgaris with abdominal tumors can not be associated where abdominal tumors do not trigger the occurrence of PV. However, abdominal tumors can increase morbidity and provide poor prognosis.

**Case Description:** A male, 46 years old, Balinese ethnic, Indonesian citizen, came to the Sanglah Hospital Emergency Room with the chief complaint of painful sores around the body, hands, and feet since 20 days ago. Initially the patient said that there were loose, watery blister filled with serous fluid, which after a few days burst and left sores. The blister start on the feet and then spread to the hands and body. The patient was diagnosed with pemphigus vulgaris and abdominal tumors by the digestive surgery doctor. The patient had never had a similar complaint before. History of applying oil, traditional ingredients and topical drugs was denied. The patient was treated mainly with methylprednisolone and showed clinical improvement.

**Conclusion:** We report a case of PV in a 46-year-old man with abdominal tumors. This case report is expected to add insight on how to diagnose PV, how to determine therapy, the wound healing process, how to treat wounds, prevent disease expansion, and the relationship between abdominal tumors and PV.

**Keywords:** Pemphigus vulgaris, paraneoplastic pemphigus, abdominal tumors.
CASE REPORT

Patient said that there were loose, watery blister filled with serous fluid, which after a few days burst and left sores. The blister start on the feet and then spread to the hands and body. The patient denied fever, cough, and runny nose. The patient also denied a similar history before.

The patient felt a lump in the abdomen and caused the stomach to enlarge since 1 year ago, then received treatment and was diagnosed with an abdominal tumor by the digestive surgery doctor. The medical history was about twenty days ago from a public health care and was diagnosed with chickenpox, so the patient was given antiviral tablets and antibiotics. The patient had never had a similar complaint before. History of applying oil, traditional ingredients and topical drugs was denied. History of previous oral medication was denied. A family history of the disease with similar complaints was denied. Work history in the family, father is a farmer and lives in a village that is quite beautiful and is said to be quite clean. In the past, the patient had an multislice computerized tomography (MSCT) scan of the thoracic axial reformat sagittal and coronal without and with contrast with the impression that there was a solid mass in the right and left paraaortic levels at Th 12-14 segment and caused mild left hydroureteronephrosis, bilateral pleural effusion with partial collapse of left lung and spondylosis in the thoracolumbar region.

On physical examination, the patient's general status was moderate and his consciousness was comos mentis. Blood pressure 110/70 mmHg, respiratory rate 20 times/minute, pulse 84 times/minute, axillary temperature 36.7°C and visual analog scale (VAS) 2/10. In general status, the head was normocephalic, on examination, both eyes did not appear anemic and no jaundice. Examination of the ears, nose, throat found no abnormalities and no enlarged lymph nodes were found in the neck. The mouth looks eroded and the tongue looks whitish. Examination of the thorax, the heart obtained a single heart sound (S1 and S2), regular, no murmurs and gallops. In the lungs, breath sounds were vesicular, no rhonchi or wheezing were found. On abdominal examination, bowel sounds were within normal limits, there was no distension, no enlargement of the liver and spleen, and a palpable mass. Warm upper and lower extremities, and pitting edema in both lower extremities. Dermatological status, location of the dextra et sinistra superior palpebral region, anterior abdominal thoracic segment, dextra et sinistra superior and inferior extremities, inguinal dextra et sinistra, lips with multiple bullae, well defined border, round shaped, scattered discretely, with a diameter of 0.5 cm x 2 cm loose wall filled with serous fluid. Multiple erosions, well defined border, geographical shape, with diameter of 0.5 cm x 1 cm – 7 cm x 16 cm, scattered discretely, partly covered with blackish brown crusts. It has mousy odor with affected body surface area (BSA) 27% (Figure 1a-1e).

The patient was differentially diagnosed with pemphigus vulgaris, paraneoplastic pemphigus, pemphigus vegetans, with other differential diagnoses being bullous pemphigoid and abdominal tumors. Blood routine results were obtained as follows: leukocytes 12.74 (4.10-11.00x10³/µL); neutrophils 78.01 (2.50-7.50x10³/µL); lymphocytes 9.87 (1.00-4.00x10³/µL); monocytes 3.50 (0.10-1.20x10³/µL); eosinophils 5.84 (0.00-0.50x10³/µL); basophils 2.79 (0.00-0.10x10³/µL); erythrocytes 5.44 (4.00-5.20x10³/µL); hemoglobin 14.99 (12.00-16.00 g/dL); hematocrit 44.50 (36.0-46.0%); platelets 447.20 (140.0-440.0x10³/µL). On clinical chemistry examination, the SGOT was 21.5 (11-27 U/L); SGPT 27.3 (11-34 U/L); BUN 24.30 (8-23 mg/dL); current blood glucose 67 (70-140 mg/dL); albumin 2.1. On electrolyte examination found sodium 120; potassium 5.94; chloride 85.

The patient was diagnosed with pemphigus vulgaris and abdominal tumor. The management given is infusion of 0.9% NaCl 30 drops per minute, methylprednisolone 125 mg injection every 24 hours intravenously, sucralfate syrup 15 milliliters every 8 hours intraoarally, nystatin drop 1 drop every 8 hours intraorally, wound dressing with NaCl 0.9% every 8 hours.
on lip lesions and eroded lesions on the body, hydrocortisone 2.5% + gentamicin 0.1% cream every 12 hours topically on eroded lesions, triamcinolone acetonide in ora base every 12 hours topically on the lip lesions, observation of vital signs, fluid balance, and progression of lesions every 8 hours, planning consult to internal medicine, digestive surgery, nutrition, and ear nose throat department.

Patients diagnosed by internal medicine doctor with hypoalbuminemia et causa suspected loss and chronic inflammation, pemphigus vegetans differential diagnose pemphigus vulgaris differential diagnose paraneoplastic pemphigus, asymptomatic chronic hyponatremia hypoosmolar hypovolemia et causa suspected loss differential diagnose low intake and received transfusion therapy of 20% (100 milliliters) albumin one flash per day up to albumin >2.5, infusion of 0.9% NaCl 20 drops per minute, general condition improvement. Patients diagnosed by digestive surgery with extrarenal abdominal tumor suspected of left ureteral infiltrating lymphoma, hydroureteronephrosis of the left kidney, bilateral pleural effusion suspected of metastatic process, hypoalbumin, hyponatremia received therapy with methylprednisolone, pro-laparotomy elective biopsy via polyclinic. The patient was diagnosed by the nutrition department with moderate malnutrition, received 2100 calories of oral nutrition therapy, 600 calories of soft food, 150 ml of msg broth every 8 hours intraorally, vitamin C 12.000 IU gargled and swallowed every 8 hours intraorally, vitamin A 10 ml of VCO + 100 ml of mineral water, 100 ml of VCO + 100 ml of mineral water, gaggled and swallowed every 8 hours intraorally, 200 ml of Entramix® every 4 hours intraorally, vitamin A 12.000 IU every 24 hours intraorally, zinc 20 mg every 24 hours intraorally, vitamin C 500 mg every 12 hours intraorally. The patient was diagnosed by the ear nose throat department with stomatitis, and received oral hygiene therapy.

On a follow-up examination 22 days after admission, the patient was found responsive to the treatment given. The patient had no complaints of new lesions, old lesions have started to heal and became dry. History of fever and pain were denied. No abnormalities of the vital signs were seen. Dermatological status show a multiple well-defined erosions, geographical in shape, discrete, with the size of 0.5 cm x 1 cm – 6.5 cm x 15 cm. Some of the lesions are covered with brown crusts. Improved mousy odour and BSA (15%) were seen. The patient had received informed consent and agreed to share the clinical picture and history for education and publication.

**DISCUSSION**

Pemphigus is a group of diseases that affect the skin and mucous membranes. This chronic autoimmune disease is histologically characterized by intra-epidermal bullae due to acantholysis (impaired adhesion bonds between cells). Epidemiologically, 80% of cases of pemphigus are PV, can affect all nations and races of women. The prevalence of pemphigus vulgaris in women is higher than that in men, namely 1.4:1. In this case, the patient was a 46-year-old male. Typical symptoms that can be found in pemphigus vulgaris are the presence of loose bullae, break easily, positive Nikolsky’s sign, and the presence of IgG against intercellular antigens in the epidermis. Nikolsky's sign can be found by means of normal-looking skin that peels off when gently pressed with the tip of the finger or by the contents of the bulla that are still intact widening when pressing. Bullae above normal skin which then develop into bullae with an erythematous base, then rupture, causing erosions which are usually accompanied by crusting and pain. Loose and easily ruptured bullae distinguish pemphigus from other chronic vesiculobullous dermatoses. The broken bullae will leave the skin peeling to form a crust. From the anamnesis it is known that the patient complained of sores around the body, hands, and feet since 20 days ago. Initially the patient said that there were watery blister that looked loose, serous fluid, which then after a few days became bursts and sores. The blister start on the feet and then spread to the hands and body. The blister are loose and break easily, if they burst they release a clear liquid.

The pathogenesis of PV is mediated by autoantibodies (autoAbs) that directly fight desmoglein 3 which is located on the cell surface of keratinocytes. The precipitating mechanism that initiates the immune response is unknown. Desmoglein 3 works as a glue that functions to attach adjacent epidermal cells through a single point of attachment called desmosome. When antibodies attack desmoglein 3 it causes cells to separate and the epidermis is detached which is known as acantholysis. Histopathology in the form of suprabasal intraepidermal bullae, acantholysis of epithelial cells, Tzanck test: acantholytic cells. Direct IF immunology: IgG and C3 intercellular antibodies. Indirect IF: IgG-type pemphigus antibody. In this case, the diagnosis was mainly based on clinical finding, while histopathological examination as a support was not carried out because it was able to distinguish pemphigus vulgaris from other types of pemphigus based on clinical finding.

The diagnosis of pemphigus vulgaris on this patient was made solely on clinical features of the patient. Supporting diagnostics such as histopathological examination, Tzanck preparation, immunofluorescence, and ELISA were not done due to the adequate information obtained from the clinical features of the patient to exclude other types of pemphigus. Patients with pemphigus vulgaris show hallmark clinical picture of thin-walled vesicobullous lesions, which will eventually rupture forming erosive lesions. In addition, pemphigus vulgaris frequently affect the mucocutaneous sites such as the ocular, oral, respiratory, and anogenital mucosa in the form of superficial blistering and persistent ulcerative lesions. Up to 90% of the patients will show oral lesions which may initially form as vesicobullous lesion which will then burst, and new lesions develop as the older lesions rupture and form ulcerative lesions.

In the case of PV, we can make a differential diagnosis with paraneoplastic pemphigus because the patient has an abdominal tumor. It can also be differentially diagnosed with pemphigus vegetans, as well as bullous pemphigoid. Paraneoplastic pemphigus (PNP) is a multiorganic autoimmune disease, usually triggered by neoplasia, mainly of lymphoproliferative origin such as chronic lymphocytic leukemia, multiple myeloma, non-Hodgkin’s lymphoma,
Castleman disease, and thymoma. PNP can be triggered by several types of neoplasia, however, approximately 84% of all patients exhibit haematological neoplasia. Non-Hodgkin’s lymphoma was the most common neoplasia with 38.6% of cases, followed by chronic lymphocytic leukemia and Castleman disease with 18.4%. Among non-haematological neoplasia, sarcoma occur in about 8.6% of cases, such as leiomysarcoma, malignant nerve sheath tumor, poorly differentiated sarcoma, reticular cell sarcoma, dendritic cell sarcoma, liposarcoma, and follicular dendritic cell sarcoma which is a tumor in the lower abdomen. The typical clinical symptom in paraneoplastic pemphigus is the presence of cancer sores that appear at the onset of symptoms, and are resistant to any therapy. These cancer sores can be in the form of ulcer that affect the oropharynx and reach the lip. In addition, there is also pseudomembranous conjunctivitis that can cause scarring of the eyes. Lesions on the skin are also polymorphic may include erythematous macules, loose wall bullae and erosions resembling pemphigus vulgaris, there are also tense bullae resembling bullous pemphigoid. The occurrence of bullae and skin lesions resembling erythema multiforme on the hands and soles of the feet can differentiate paraneoplastic pemphigus from pemphigus vulgaris. Pemphigus vegetans is a rare type of pemphigus. It is characterized by the appearance of loose bullae that become eroded lesions and then papillomatous proliferations especially in the folds of the body and scalp and face. The initial lesions are pustules, more numerous than vesicles, and progress to vegetative plaques. Bullous pemphigoid is a type of pemphigus whose cause is thought to be autoimmune. Usually occurs in old age, there is no itching, and found bullae in the walls of tension. The predilection is on the abdomen, hand flexors, inguinal, and medial leg. Oral mucosal abnormalities ranged from 10-40% and Nikolsky’s sign was absent.1,2,10

Abdominal tumors constitute one third of all malignant tumors. Abdominal tumors are solid masses of varying thickness caused by abnormal growth and transformation of body cells. Clinical symptoms of abdominal tumors are difficult to detect due to the nature of the abdominal cavity which is quite flexible and loose. Symptoms will be detected after the stomach looks bloated and hardened, has swelling, difficulty defecating, pain, no appetite, nausea, lethargy, bleeding, and drastic weight loss. The causes of abdominal tumors can be carcinogenic materials, hormones, lifestyle, parasitic infections, trauma, and genetics.9,10

Choosing the right dressing is important in accelerating the wound healing process, where the basic principle is that a wound with a moist environment will heal faster than a dry wound.12,13 This moist environment can be achieved by applying dressings that are able to maintain wound moisture, including films, hydrogels, hydrocolloids, foams, alginate and hydrofibers. The ideal dressing requirements, among others, meet the general characteristics (easy to apply, aesthetically acceptable, affordable cost, easy to store, non-allergenic), facilitate the healing process (capable of maintaining a moist environment, not causing trauma or maceration at the wound edges, able to withstand heat, as a means of gas exchange), and minimize the risk of infection (capable of cleaning necrotic tissue, absorbing exudate, minimizing external contamination). Each dressing is used in different clinical applications and has its own advantages and disadvantages.14,15

The four main goals of wound care are (1) infection prevention, (2) maintenance of a moist environment, (3) wound protection, and (4) minimizing scar formation. Small blisters should be left intact, to prevent secondary infection. However, large blisters should be punctured and aspirated with a large sterile needle (size 18), to keep the surface of the blister in place for wound protection. Initial wound washing can be done with physiological saline or soap/antiseptic twice daily. Next, you can apply a soft emollient ointment, such as 50% white soft paraffin and 50% liquid paraffin, which is applied directly to the wound bed, or apply the ointment to the dressing. If the wound is wet, the dressing will absorb the excess exudate. If the wound is dry, the dressing should provide moisture to the wound bed.7,16,17

The first agent to be considered for pulse dose therapy in pemphigus vulgaris is methylprednisolone. Furthermore, the combination of dexamethasone and cyclophosphamide which has been introduced as pulse dose therapy in several countries can also be used, because of its lower cost and easier availability. However, this combination may be associated with a high incidence of relapse and other major adverse events particularly associated with long-term administration of cyclophosphamide.7,18

In this case, the patient received therapy in the form of steroid injection, namely methylprednisolone. Besides of controlling the disease by healing the lesions, the real challenge of treating pemphigus vulgaris is to achieve rapid remission with minimum flares and morbidity associated with treatment agents. After 22 days of observation, the patient did not show signs of relapses and adverse events of using immunosuppressant agent. Adverse events of corticosteroid administration include alterations in musculoskeletal (myopathy and osteonecrosis), metabolic, endocrinial, infections, cardiovascular, ophthalmologic, gastrointestinal, and neuropsychiatric systems. Corticosteroid may alter the integumentary system in particular showing ecchymosis, skin thinning, atrophy, acne, hirsutism, facial erythema, striae, impaired wound healing, thinning of hair, and perioral dermatitis,19 in which were not seen in this patient.

Tetracycline, mycophenolate mofetil, rituximab, and azathioprine can be used as steroid sparing agents. Immunosuppressive drugs are given in addition to or in lieu of steroid therapy to reduce the steroid dose required and thereby avoid some of the unwanted side effects of corticosteroid therapy. For this reason, these drugs are sometimes referred to as steroid sparing agents.18,20

In one study, sixteen patients were given nicotinamide 1.5 g and tetracycline 2 g daily. In 12 patients, no systemic steroids were administered and only three cleared and three improved. Of the four patients given additional prednisolone, there was clearance in one patient, partial improvement in two and no response in the other. Tetracyclines with or without nicotinamide may be considered an
adjunct treatment, perhaps in milder cases of pemphigus vulgaris. In this case, the patient's had poor prognosis due to comorbidities along with the PV.

CONCLUSION
A case of pemphigus vulgaris has been reported in a 46-year-old male patient with comorbid abdominal tumors. The diagnosis is made based on history, physical examination, and investigations. Management of PV patients with multiple comorbidities requires collaboration with other specialists.

CONFLICT OF INTEREST
No conflict of interest.

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AUTHOR CONTRIBUTIONS
Authors and co-author responsible in taking care, and follow-up the patient daily, manuscript preparation, and publication.

REFERENCES